Ocular hypertelorism is a congenital craniofacial malformation characterized by excessive distance between the eyes. It is one of the most expressive deformities of the human face. The aim of this study was to discuss the clinical aspects associated with the diagnosis, planning, and treatment of an orthodontic patient with ocular hypertelorism.

DIAGNOSIS AND ETIOLOGY

An 8-year-old girl sought treatment at the clinic of the Specialization Course in Orthodontics of the University of Itauna in Brazil. She exhibited facial asymmetry, ocular hypertelorism, increased lower third of the face, convex profile, acute nasolabial angle, hypotonia of the lower lip, and absence of lip seal. The oral examination showed that she was in the mixed dentition with an Angle Class II malocclusion. The overjet was 13 mm, and she had an anterior open bite and a bilateral posterior crossbite. The maxillary arch was atresic and crowded, and the mandibular arch had a diminished perimeter (Figs 1 and 2). The radiographic examination showed the following: agenesis of the maxillary left second premolar; a supernumerary lateral incisor and a supernumerary premolar on the right side; history of trauma, endodontic treatment, and resorption of the maxillary left central incisor; and root resorption of the maxillary right first molar caused by the second molar (Figs 3 and 4). The cephalometric evaluation showed that most dental and skeletal parameters were outside the standards of normality, and there was a tendency toward vertical growth (Fig 5). The parents reported that the patient had undergone surgery at the Lucile Salter Packard Children’s Hospital in Stanford, California, for the correction of hypertelorism 2 years earlier.

Hypertelorism can be caused by numerous congenital defects in the development of the face and cranium or result from trauma. It might also be confused with many types of pseudohypertelorism, in which there is broadening of the interorbital distance because of an abnormality in the width of the bone between the sockets. True hypertelorism is associated with diverse...
congenital deformities, but there are 4 main groups: pure hypertelorism (Greig’s syndrome), characterized by broadening of the nose and eyes, short and swollen fingers, heavy eyelids, and lateral exotropia; hypertelorism associated with frontal encephalic hernia or excess skin along the midline; hypertelorism associated with severe lateral facial clefts and clefts of the lip and palate; and acrocephalosyndactyly hypertelorism (Apert’s syndrome), a syndrome of trisomy 21 and other defects that result from autosomal dominant genes. In the case described here, the patient had no neurologic impairment. She had intelligence proportional to girls of her age and exhibited concern and considerable interest about improving her facial appearance.

TREATMENT OBJECTIVES
The treatment objectives were to extract the supernumerary teeth, eliminate the crowding in the maxillary and mandibular arches, correct the posterior crossbite, eliminate the anterior open bite, promote passive lip seal, and achieve a Class I skeletal and dental pattern. A further aim was to improve the esthetics of the nose through rhinoplasty.

TREATMENT ALTERNATIVES
To achieve the treatment objectives, 2 alternatives were considered: surgical correction of the Class II malocclusion and orthodontic camouflage through dental compensations. Surgical correction could involve moving the maxilla backward or advancing the mandible. Because the patient had already had an invasive surgical procedure at 6 years of age and was about to undergo rhinoplasty as well, the decision was made for the more conservative treatment: orthodontic camouflage.

The treatment plan involved the following steps: (1) maxillary expansion with a Haas appliance,
a transpalatine bar, and an oblique headgear; (2) extraction of the maxillary right incisor and first molar because of root resportion; and (3) rhinoplasty. An orthopedic approach with an activator was discarded, since the patient exhibited abnormalities in her craniofacial growth pattern. However, the oblique headgear was indicated for anchorage and restricting forward movement of the maxilla, which had an SNA angle of 85°. In the
place of the extracted first molar, the decision was made to close the space by using the supernumerary premolar. The maxillary right lateral incisor was transformed with composite resin to replace the central incisor. There was a possibility that the extracted teeth could be replaced with implants or even fixed dentures. Once again, however, the decision was made for a more conservative approach, since the patient and her parents were motivated and receptive to the orthodontic treatment.

TREATMENT PROGRESS

Treatment began with the placement of the Haas expander, which was activated twice a day (a quarter turn in the morning and at night). After 2 weeks of activation, the expected orthopedic effect had not been achieved (Fig 6). However, the appliance was maintained as a retainer for 4 months, considering that the expansion was purely dental. The supernumerary lateral incisor, maxillary right central incisor, maxillary right first
molar, and maxillary left second deciduous molar were extracted. The palatal bar and oblique headgear were installed. Alignment and leveling of the arches was initiated, followed by mechanical retraction and the use of Class II rubber bands. The lateral incisor was then restored as a central incisor with composite resin (Fig 7).

**TREATMENT RESULTS**

The functional results achieved were satisfactory considering the limitations of the case (Figs 8 and 9). The patient exhibited bilateral Class II canines, with 3 premolars on the maxillary right side. There was also an improvement in overjet, which was reduced by 13 mm to a normal relationship. The overbite and crossbite were corrected. However, the profile was slightly concave because of an inadequate relationship between the lips. The cephalometric evaluation showed a few skeletal abnormalities and significant dental movement, represented above all by the retraction and extrusion of the maxillary incisors (Figs 10 and 11).

The patient had considerable improvement in her self-esteem, and the parents were happy with the results.

**DISCUSSION**

Surgical correction of hypertelorism involves significantly invasive techniques. The danger of injury to the brain or optic nerve, meningitis, paralysis of the extraocular muscles, and postoperative diplopia are some reasons that surgeons hesitate to perform such a procedure. Without correction, these children, although often of normal intelligence, are condemned to live isolated from society. However, recent advances have contributed to better surgical results. The facial bipartition technique is considered a considerable advance in achieving a more natural appearance for the correction of hypertelorism, especially in those operated on after the peak growth spurt (14-15 years of age). In patients operated on during the first decade of life (2-10 years), significant relapse is often observed of the anatomic positions achieved during surgery. Our patient underwent the surgical procedure before her peak growth spurt.
Nonetheless, no significant relapse was observed 4 years after the surgery.

Another aspect related to the orthodontic correction is the possible repercussions to the growth of the craniofacial structures directly involved in the orthodontic treatment, such as the growth and displacement of the maxilla. A detailed analysis of 4 patients based on Ricketts’ long-term growth forecast showed a reduction in growth in the region of the anterior nasal spine only in the posteroanterior direction.4 This is not necessarily due to the absence of the nasal septum. These patients have such severe deformities, and the surgical procedure is so extensive that many factors can be responsible for deficient premaxillary growth.4 Another study showed that resection of the ethmoid bone located between the orbit and the medialization of the orbital skeleton through the intracranial approach modifies the exaggerated interorbital distance but does not correct the vertical shortness at the midline of the face. Moreover, this procedure interferes with the sagittal growth of the maxilla, possibly resulting from the horizontal osteotomy across the maxilla. The medial rotation of the 2 halves of the face in the intracranial approach or the subcranial approach simultaneously corrects the orbital hypertelorism and elongates the nose and the central segment of the face.4 In our patient, there was a limitation to the anterior displacement of the maxilla, possibly associated with the action of the headgear and the surgical procedure. This effect directly contributed to the correction of the Class II malocclusion. On the other hand, the
horizontal osteotomy across the maxillae performed pre-
maturely might have led to the unsuccessful maxillary expansion.

Our patient also had anomalies in dental develop-
ment (agenesis and supernumeraries), thereby suggest-
ing a genetic influence, although there were no reports
of any family members with any condition associated
with hypertelorism.

The different limitations imposed by bone and dental
aspects associated with hypertelorism impeded achiev-
ing a standard of excellence considering the occlusal ob-
jectives of the treatment. However, the patient and her
parents were happy with the results and reported that
the orthodontic treatment had a significant effect on
her quality of life.

CONCLUSIONS

The treatment of patients with ocular hypertelorism
imposes different limitations and is a difficult challenge
for orthodontists and surgeons alike. A multi-
disciplinary approach focusing on familial, psychologi-
cal, surgical, and orthodontic aspects could contribute
toward the success of treatment.

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